The Surgical Management of Infants and Children With Ambiguous Genitalia

Lessons Learned From 25 Years

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Over a 25-year period, 91 children with ambiguous genitalia have received surgical management. Female sex assignment was made for 79. Of these, 60 patients underwent extensive clitoral reconstruction consonant with the female assignment. Forty-two patients had vaginal reconstruction. Factors relating to success include: (1) prompt and appropriate sex assignment; (2) early and accurate diagnosis; (3) conservative reconstruction of the clitoris at an early age (less than 1 year); and (4) choice of vaginal reconstruction based on the severity of the malformation. Long-term follow-up demonstrates satisfactory anatomic and functional results when clitoral surgery alone was required. Functional results for patients with extensive vaginal reconstruction have been compromised. Physicians caring for children with congenital intersexual anomalies can expect to encounter a wide spectrum of anatomic and physiologic derangements. Cosmetic appearance alone is an inadequate measure of success because endocrinologic, social, psychological, and sexual factors must be blended into comprehensive evaluation of these patients. The management plan must be flexible and individualized, incorporating long-term follow-up to adulthood.

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AMBIGUOUS GENITALIA, WHICH herald some variant of intersexual malformation, are now recognized immediately in the newborn nursery. The tragedies of yesteryear when an adolescent girl would suddenly grow facial hair or a teenaged boy would develop breasts unexpectedly rarely occur. Diagnostic tools are available for use in the first days of life, which permit assessment of biochemical derangements, determination of genotypic pattern, and definition of internal genital anatomy. Based on these observations, an accurate and appropriate sex assignment can be issued with confidence and dispatch. This decisive step is urgent, minimizing the psychosocial impact of a parent rendering an equivocal or evasive answer to the age-old question from loved ones, as to the sex of the new baby. Once the sex of rearing is firmly established, a plan for the required surgical correction and its timing assumes paramount importance.

From the patients reported here, certain principles have been derived that have been found helpful in managing these complex clinical problems over the last 25 years.

Materials and Methods

Ninety-one children with ambiguous genitalia have been cared for since 1966 at our institution. The follow-up ranges from 1 to 24 years, with a mean of 8 years. The clinical records of all patients were analyzed. Additionally, many patients have been seen regularly, others sporadically, and, sadly, some have been lost to follow-up. The results of the last examination were reviewed, and when available the older patients were brought back for full evaluation. Outcome was evaluated as to appearance, and of special importance, older patients were subjected to assessment of psychosocial and sexual function. A single surgeon provided the unified, continuous surgical care of these children. The comprehensive support that these patients need was provided by nurses, neonatologists, pediatricians, endocrinologists, psychologists, and parents.

Diagnosis and Sex Assignment

In the newborn infant, a precise diagnosis is achieved by combining the results of the physical examination with the laboratory evaluation. The location of the gonads, the adequacy of the phallus, the size and location of a vaginal orifice, if present, are assessed (Fig. 1). A rectal examination provides information concerning the presence of a uterus or prostate gland. Introduction of x-ray contrast

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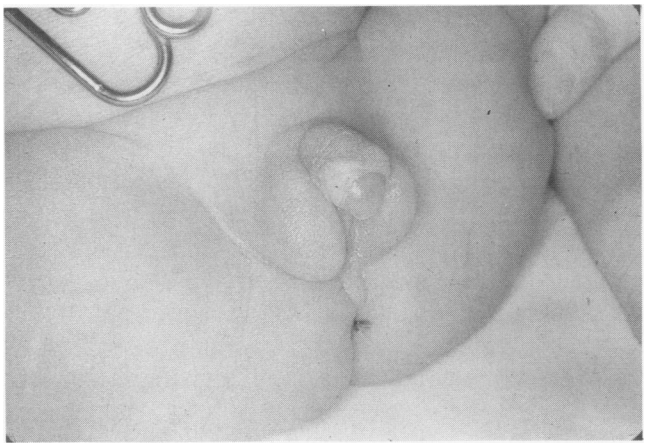


FIG. 1. Ambiguous genitalia as seen in the newborn.

into a urogenital sinus outlines the internal urethral and vaginal anatomy (Fig. 2). Ultrasonography supplies useful information concerning the internal pelvic organs.

An expeditious laboratory evaluation is carried out to determine the specific genotype and pathophysiologic cause for each child. The chromosomal makeup is determined from the presence of chromatin on a buccal smear and blood cell karyotype analysis. Serum electrolytes, steroid levels, and urine metabolites pinpoint errors in steroid biosynthesis. Although many sophisticated tests are becoming available, the absolute initial requirement is a rapid determination of the genotype, and identification of the potentially life-threatening salt-losing adrenogenital syndrome.

When all information from the physical examination, the laboratory, the radiologic/imaging studies, and, in rare instances, exploratory laparotomy is at hand, the sex of rearing can be assigned with confidence. The male role is naturally elected for all genotypical males born with hypospadias and undescended testicles. Male pseudohermaphrodites, including some with testicular femininizing syndrome, an occasional true hermaphrodite, and some infants with gonadal dysgenesis may be successfully reared as males, if, and only if, they are born with a male-sized penis. A small subgroup of male (XY genotype) patients exists who are born with hypoplasia of the penis. These patients suffer from interruption of the synthesis of dihydrotestosterone (5-alpha reductase deficiency) or other poorly understood causes of phallic inadequacy. Because the phallus is so small as to resemble a clitoris, these patients are unsuited for the masculine role and must be raised as girls.

All of the female patients with congenital adrenal hyperplasia as well as females with clitoromegaly from other causes should be reared as girls. These patients constitute approximately 90% of the entire cohort with intersexual malformations, and most require some form of reductive surgery for the clitoris. In addition, most will need vaginal reconstruction.

In practical terms, regardless of the genotype, most children with ambiguous genitalia are best suited for the female role. In the highly selected cases to be brought up as males, the crucial determinant is the size of the phallus, that is, whether it is adequate to support a male sex assignment. If not, female sex assignment is made and planning for total rehabilitation is outlined. This program of comprehensive staged surgical reconstruction, and in many patients, a lifetime of exogenous medication is thoroughly discussed and repeatedly explained to the parents. Such discussion must be regularly revisited.

Male Reconstruction

Fewer than 10% of patients born with the intersex anomalies will be directed to a male sex assignment. These male patients are culled from the group of male pseudohermaphrodites, or occasionally a child with mixed gonadal dysgenesis, or more rarely a true hermaphrodite. The surgical requirement for male sex of rearing is the

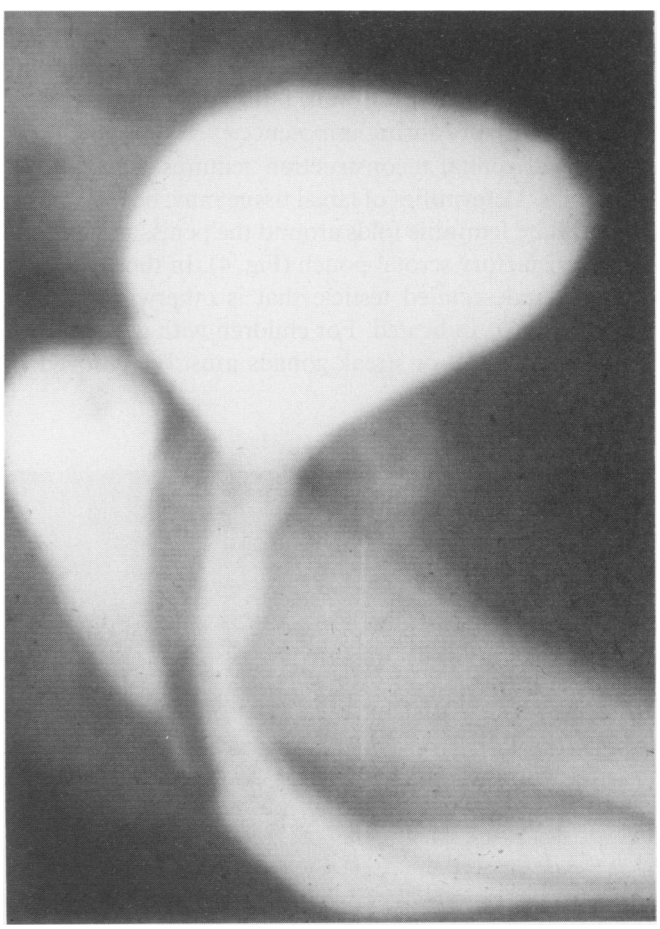


FIG. 2. X-ray study with contrast filling the urethra and bladder, showing distal vaginal atresia and urethrovaginal fistula (type C).

presence of a sufficiently large phallus to function as a male urinary conduit, to offer a satisfactory appearance when compared with peers, and to function satisfactorily for sexual activity (Fig. 3).

Of those patients most appropriate for a male sex assignment, the least complicated are those with XY genotype who happen to be born with one or both testes undescended, and have an associated hypospadias. Physical examination of such newborn infants discloses a confusing external genital presentation, which requires the standard workup. It is sometimes necessary to perform an exploratory laparotomy to obtain gonadal tissue for confirmatory histology and to exclude Müllerian remnants. Such patients engender the least social stigmata and can be set on their life path as whole and healthy males with two standard operations: orchidopexy and hypospadias repair.

In those more complex patients to be raised as male, notably a rare true hermaphrodite, or more commonly those pseudohermaphrodites with a normal-sized penis, several orderly steps lead to reconstruction. If Müllerian duct structures persist in the pelvis, they are surgically removed. These structures may include an ovotestis, Fallopian tube, uterus, and proximal vagina; the distal vagina should not be dissected for fear of injuring the adjacent nerves. This maneuver leaves a persistent diverticulum on the posterior urethral wall, but is to be favored over the possibility of causing impotence.

External genital reconstruction requires repair of hypospadias. Deformities of labial tissue must be rearranged to eliminate feminine folds around the penis, and to fashion a satisfactory scrotal pouch (Fig. 4). In those patients with an undescended testicle that is otherwise normal, orchidopexy is indicated. For children with gonadal dysgenesis, imperfect or streak gonads must be removed to avoid malignant degeneration. Testicular prostheses are recommended as the youngsters reach adolescence.

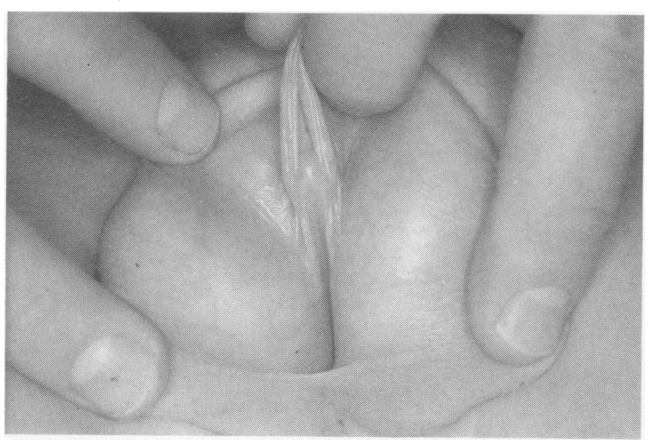


FIG. 3. Genotypic male with end organ defect. This inadequate phallus determines female sex of rearing. Phallus length is compared with published "normal" values.[41]

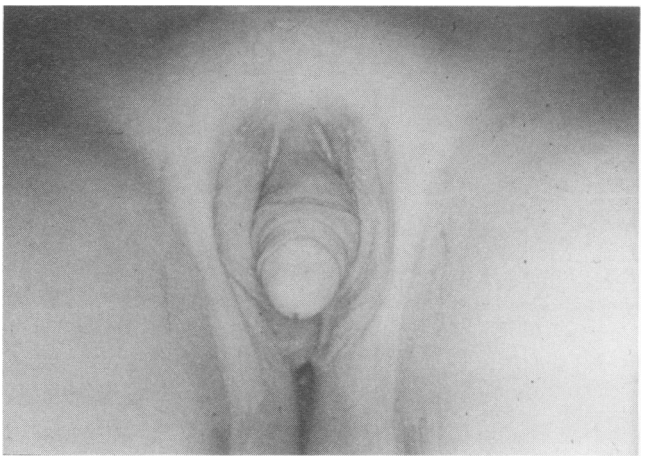


FIG. 4. Male pseudohermaphrodite with penis adequate for male sex rearing. Transposition of labia to create scrotum to maximize masculine appearance is essential.

Female Reconstruction

Clitoral Reconstruction

Most patients with intersexual malformations are best suited for female sex assignment. These infants have been derived from a wide range of diagnoses, and most require surgical correction of an enlarged clitoris (Table 1). It is the clitoral-sparing recession operation, devised in our institution, which we have used in the series of patients reported here.[1] The basic tenets of the operation, described in detail in previous reports, are seen in Figure 5. These are: (1) removal of the excess skin over the enlarged clitoris; (2) dissection of the shaft with preservation of the neurovascular bundle; (3) division of the suspensory ligament; (4) protection of the underlying urethra; (5) seating the clitoris in a retrograde manner beneath the pubis; and

TABLE 1. *Diagnoses of Patients With Ambiguous Genitalia at Children's Hospital, Washington, DC, 1966–1991*

Diagnosis	No.
Congenital adrenal hyperplasia	46
Maternal ingestion of hormones	9
True hermaphrodite	4
Male pseudohermaphrodite	7
Idiopathic	5
Gonadal dysgenesis	4
Congenital absence of the penis	2
Simple XY hypospadias with undescended testicles	6
Cloacal anomalies	6
Vaginal atresia	1
Mosaic XO–XY	1
Total	91

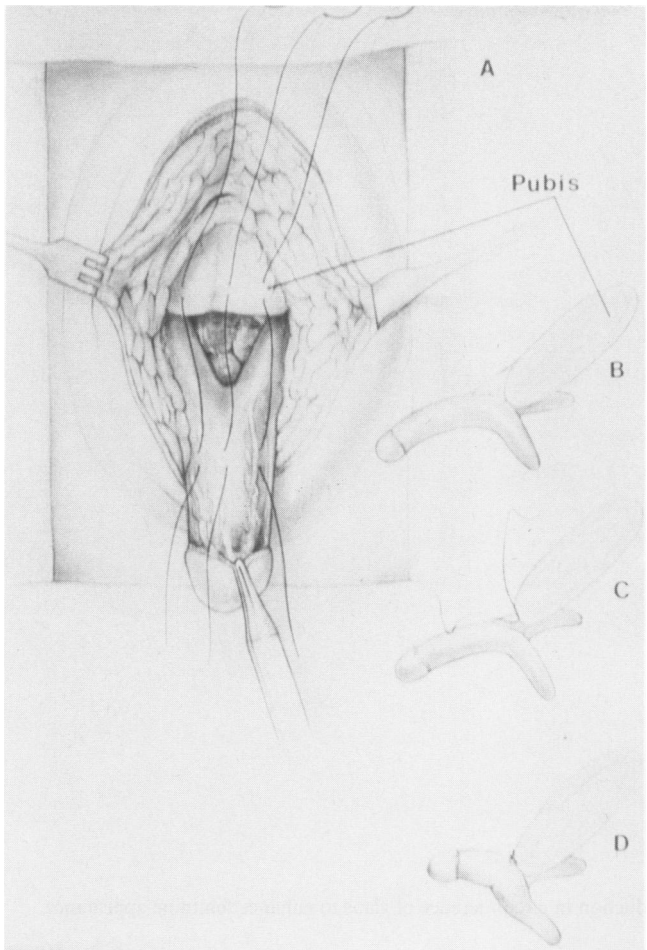


FIG. 5. Basic diagram of elements of clitoral-sparing recession. Note preservation of the corpora with recession to the undersurface of the pubis.

(6) anchoring the clitoris in the new position with sutures taken in Buck's investing fascia, which are then affixed to the periosteum of the undersurface of the pubis. The surrounding fatty tissue is then brought together in the midline to recreate the mons veneris, which further minimizes the visible part of the clitoris (Fig. 6). For some patients in whom the circumference of the glans is so large as to be disfiguring, we have developed a reduction glansplasty. The anterior wedge excision results in a more delicate feminine glans clitoris (Fig. 7).

Vaginal Reconstruction

In these patients, four basic operations have been used, depending on the degree of vaginal malformation. Cystoscopy and contrast studies are useful adjuncts to define the anatomy. We currently categorize the vaginal anomalies as follows:

Type A. Simple labial fusion
Type B. Distal urogenital sinus formation
Type C. Distal vaginal atresia with proximal urethrovaginal fistula
Type D. Complete absence of the vagina

Each of these deformities requires a different reconstructive approach. The complexity of the surgery increases with the severity of the vaginal malformation.

For type A (simple labial fusion), a midline division of the labia shows a normal hymeneal ring and a normal or near-normal urethral meatus. After division of the labia, the skin and mucosal layers are approximated to assure that the introitus remains open, creating a normal appearance.

For type B (distal urogenital sinus formation), a more extensive separation of the fused tissue is required. It is necessary to develop a posterior pedicle flap from the labioscrotal tissue, which then is sutured to the vagina, creating a satisfactory opening for the urinary and genital orifices.

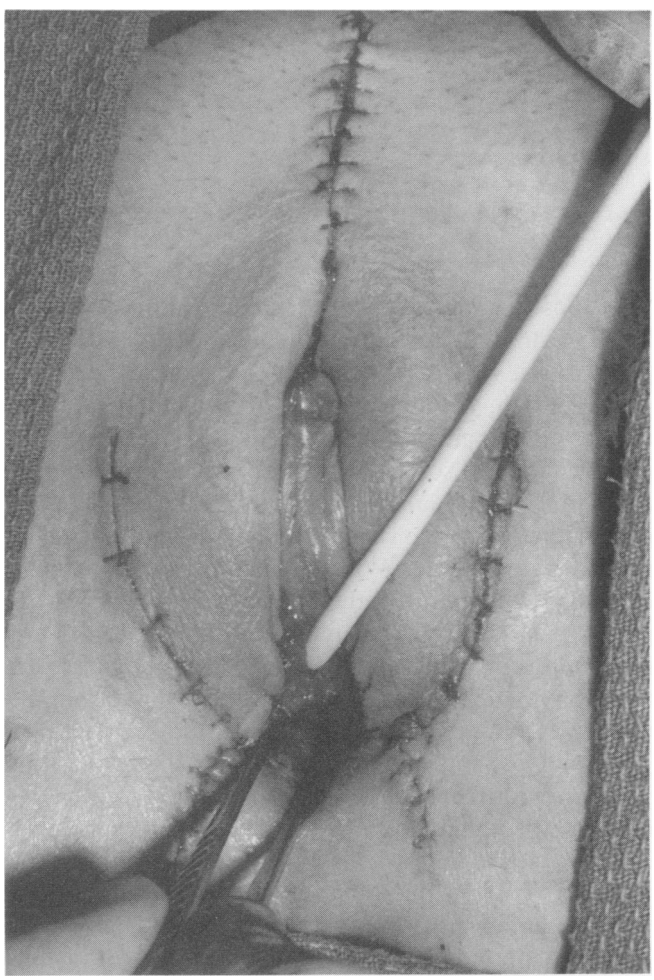


FIG. 6. Appearance immediately after surgery, showing recession clitoroplasty and vaginal reconstruction. Note reconstruction of introitus using labioscrotal folds.

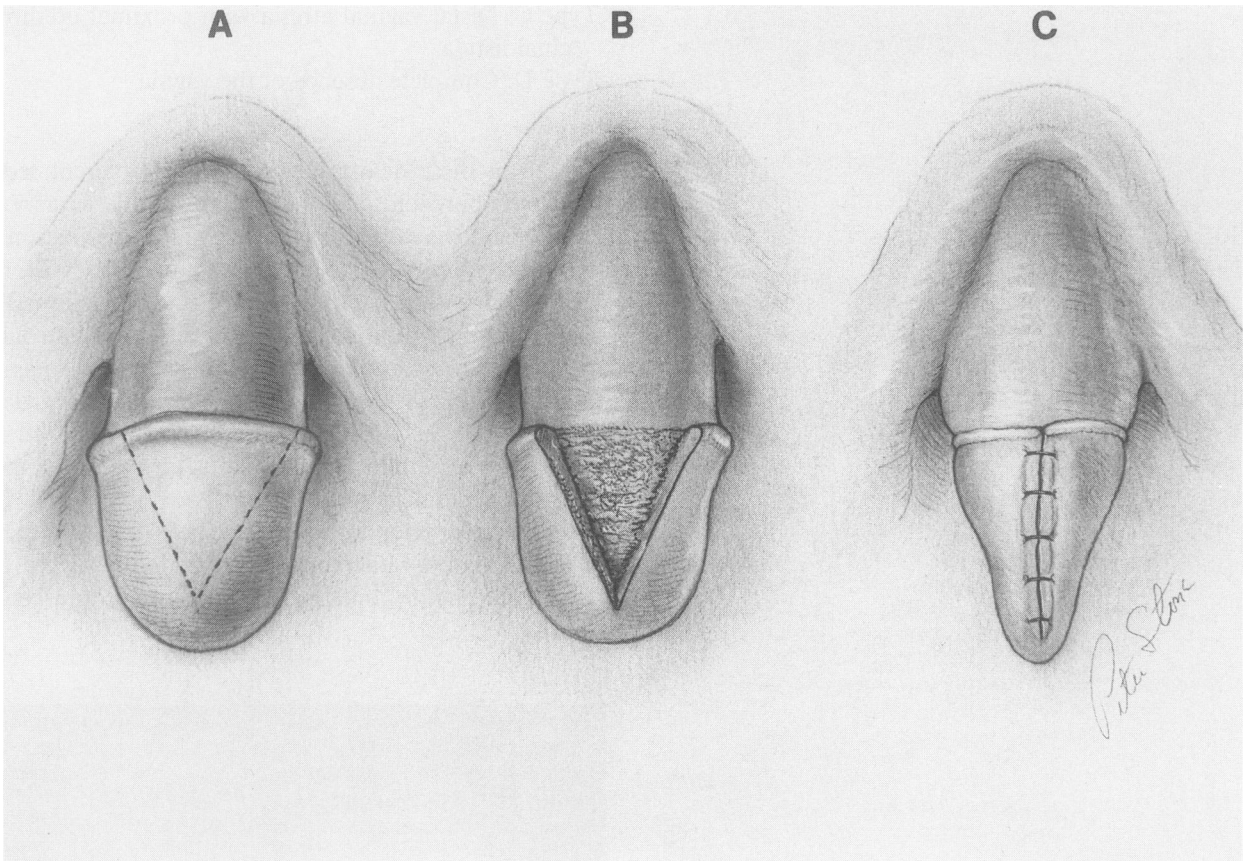


FIG. 7. Operative diagram showing glansplasty for children requiring reduction in circumference of glans to enhance feminine appearance.

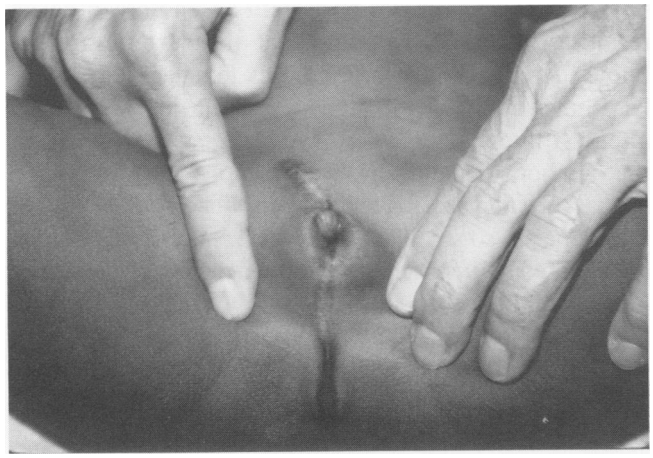
In those patients with type C anomaly (distal vaginal atresia with proximal urethrovaginal fistula), we have performed a vaginal pullthrough operation using a perineal approach. With the patient in the lithotomy position, a series of flaps are outlined and elevated. The dissection is carried into the soft tissue beneath the urethra. Hendren and Crawford have recommended the insertion of a Fogarty catheter into the vagina through the urethra and the urethrovaginal fistula.[2] With the Fogarty balloon partially expanded, recognition of the vagina is simplified. The vagina is separated from the urethra, and the urethral defect is closed over a catheter. The pedicle flaps of labioscrotal tissue are then sewn to the vagina to separate it from the urethra and to permit exteriorization of the vagina.

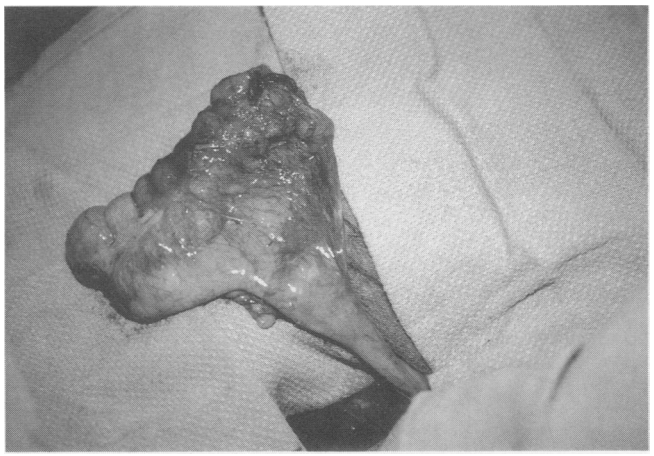
In some patients, the circumference of the vagina at its junction with the external pedicle flaps will become inadequate or frankly stenotic. In such circumstances, we have resorted to the use of large well-vascularized pedicle flaps derived from the inner aspect of the thigh. These flaps are outlined just below the skin crease between the leg and the groin. The narrow vagina is incised longitudinally from outside, well inward to the midvagina. The elevated flap is drawn into the defect and sutured, thus widening the circumference of the vagina. When additional width is required, bilateral flaps are used.

Patients requiring complete vaginal replacement (type D) have thus far been encountered only rarely. As we have followed our patients into adolescence and now adulthood, however, it is becoming apparent that more patients will require total vaginal reconstruction. We have used a number of different methods to reconstruct the vagina, including chronic dilation, perineal flaps, pedicle flaps from the inner thigh, and molded stent skin grafts. None of these methods has proven fully satisfactory, and therefore we have recently embarked on a program of vaginal reconstruction/replacement using a sleeve of transplanted colon. The blood supply of the colon permits it to be mobilized and easily transposed into the pelvis. Perineal dissection allows the colovaginal transplant to be brought into the proper position anterior to the rectum and sutured to the surrounding labioscrotal tissue, which has been fashioned into an introitus (Fig. 8).

Results

Ninety-one patients were evaluated with ambiguous genitalia. In the first decade, approximately 30% were not





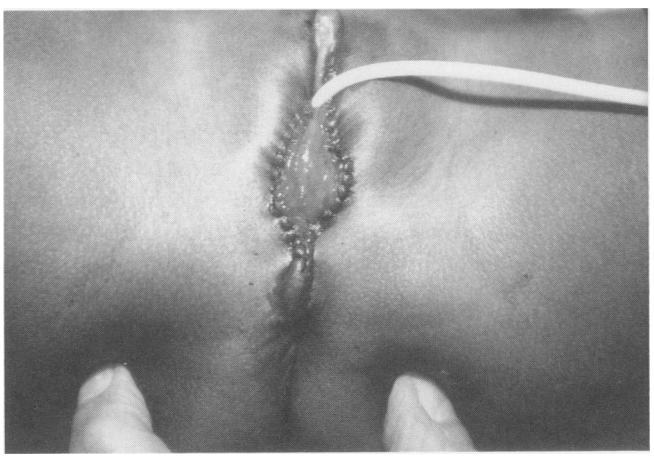


FIG. 8. (A, top left) Patient with stenosis of a vaginal pull-through requiring colon vaginoplasty. (B, top right) Colon sleeve on vascular pedicle prepared for placement in pelvis. (C, bottom left) Postoperative appearance of colon neovagina of same patient.

recognized as newborns. In the last decade, all patients were recognized in the newborn period. Of the 91 patients, 12 were managed as males. Seventy-nine received female sex assignment and were raised as females. Although most required surgical intervention, 10 females were followed without operative management.

Of the 79 female children, 60 had clitoral surgery. In the very early days of our experience, clitorectomy was the favored operation; since 1966, however, clitoral-sparing recession has been employed in most patients (Table 2). Forty-two children had vaginal surgery (Table 3).

Appearance

The external appearance of virtually all of the patients with female sexual assignment is judged as satisfactory at present (Figs. 9, 10). A significant number of children required secondary operations such as a second clitoroplasty (26%) or glansplasty (15%) to achieve satisfactory cosmetic appearance.

TABLE 2. *Operations for Clitoral Reconstruction*

Procedure	No.
Clitoral recession	53
Clitorectomy	7
Total	60
Revision	16
Second revision	2
Glansplasty	9

Function

At this time, appraisal of function is incomplete; however, 20 patients were available for long-term follow-up into adulthood. Twelve had clitoral surgery alone. Ten of these show highly satisfactory social, psychological, and sexual function. One patient demonstrates partial success but has pain with orgasm, and one has dyspareunia. Of

TABLE 3. *Vaginal Reconstruction*

Procedure	No.
Simple introitoplasty	
Incision and opening	10
Labial excision	2
Posterior flap	7
Pullthrough	14
Skin graft	3
Thigh pedicle	4
Colon	2
Total	42

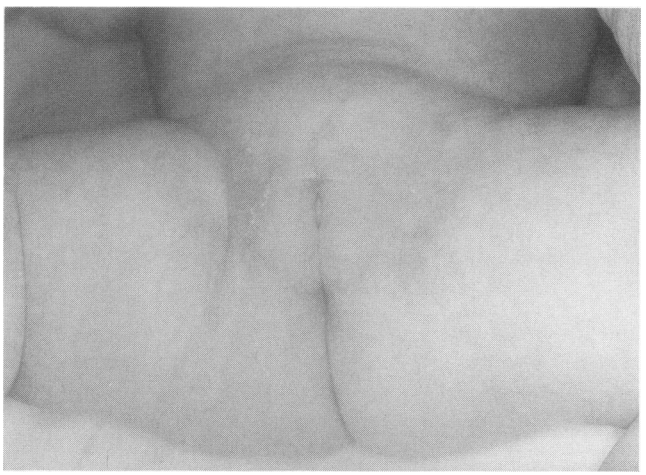


FIG. 9. Child 1 year after clitoral recession showing satisfactory feminine contour.

the remaining eight adult patients who underwent extensive vaginal as well as clitoral surgery, most have achieved acceptable results in social and psychological adjustments, but sexual function has ranged from satisfactory to poor. Two of the four patients with thigh flaps continue to have problems with vaginal size. Eight of the 14 with a vaginal pullthrough (type C) developed severe stenosis requiring secondary operations; additionally, three patients have been discovered with uncomfortable growth of hair at the introitus.

Discussion

The patients selected for the male role constitute slightly less than 10% of all infants born within the spectrum of diagnoses encompassing the intersex syndrome. As with all such patients, each requires precise and prolonged follow-up of their appearance, of their function, and for psychosexual adjustment. As part of a program of ongoing counseling, these young males must come to understand the abnormality, and when appropriate, the absence of sex glands, and the need for exogenous hormone support. Further, with some exceptions in the group of males born with the less complicated variant of hypospadias and undescended testicle or testicles, most of these children will be sterile. For all of our patients, males and females who will be sterile, it has been our practice to instruct the parents to eliminate from their conversation, and thus from their child's thinking, all traditional references to genetically transmitted characteristics, for example, "He has his mother's eyes," "He has a big nose like his father," or "He gets his stubborn streak (as an inherited trait) from his uncle John." Instead, because the future family constellation of sterile adults will probably include adopted children, the emphasis must be on the paramount importance of love between family members, and how that emotion is the key to family bonding.

With respect to males born with an end-organ defect resulting in penile hypoplasia, at the current state of knowledge, they must be raised as females. Experience has shown that the most heartbreaking maladjustment attends those patients who have been raised as males in the vain hope that the penis will grow to a more masculine appearance and size "at a later date." The phallus of such individuals is so small as to resemble a clitoris; thus, they are doomed to life as a male without a penis. We, as others, have concluded that, until such time as enzymatic synthesis can produce adequate and predictable growth of the penis in these infants, they must be raised as females. This assignment is made contradictory to the patient's XY genotype in the full awareness that exogenous hormone therapy and extensive surgery for vaginal replacement will be necessary. However, the required comprehensive rehabilitation program is preferable to a male life without a penis. Because the clitoral-like phallus is properly innervated, orgasm and sexual function in the female role is theoretically achievable, although none of the pa-

FIG. 10. Appearance of a 19-year-old patient whose clitoral recession was performed at 5 years of age. Note the accommodation of genital components.

tients available for study in this series have been followed into adult life.

Males born with congenital absence of the penis are extremely rare; two such patients have been encountered in this group over the 25-year study period (Fig. 11). Surgical rehabilitation is achieved by performing orchiectomy because, as in patients with penile hypoplasia, female rearing is the only reasonable course. A female distal urethra is constructed, positioning it appropriately in the introitus, which is fashioned from the skin of the normal scrotal pouch. Colovaginoplasty can be performed if the patient desires this, but ultimate adjustment is marginal at best, because all erotic tactile stimulation is congenitally lacking.

The existence of a normal-sized penis in patients assigned a male role is comforting to the parents even when hypospadias is present. Conversely, female babies born with an ungainly masculine enlargement of the clitoris evoke grave concern in their parents. Our experience suggests that once the female sex of rearing is made, the parents are usually calmed by a full explanation of the child's pathophysiology, and the promise of major corrective surgery within 3 to 6 months. This age has seemed ideal for the clitoral-sparing recession operation in our hands. The babies who require daily medication, as in the salt-losing adrenogenital syndrome, are stabilized by 3 months of age, and the infant's size at 3 months is more conducive to the requirements of the clitoral operation than that of a newborn infant.

There are two major approaches to plastic reduction of the enlarged clitoris, and each has strong support. Resection of all but the tip of the glans clitoris with preservation of the nerve and blood supply was suggested in early work by Goodwin,[3] and has been championed by others.[3-8] The cosmetic effect is excellent. Late studies with assessment of sexual gratification, orgasm, and general psychologic adjustment are unavailable for these patients and remain in question. Burying the enlarged clitoris by drawing surrounding fat over it was suggested by Lattimer in 1959.[9] This was the first effort to preserve all of the clitoris, as opposed to complete corporal resection.[10] In 1966 at our institution a procedure was developed that preserves all of the clitoris, and after full dissection, permits its recession beneath the central pubis with preservation of all nerves, blood supply, and corporal bodies, allowing erection without cosmetic offense.[1] Over the years, this operation has worked well, but has required a second procedure in a number of children, a third in several patients, and a glansplasty in others. Conservation of all of the clitoris, including its erectile function, would seem to provide the best chance for the patient to achieve normal orgasm in adult life. In fact, only the total clitoris-sparing recession operation has had any long-term assessment for sexual function in children who have grown to adulthood.[11-13] Results from these rather rudimentary studies are so encouraging as to make a strong case for retaining the entire clitoral organ.

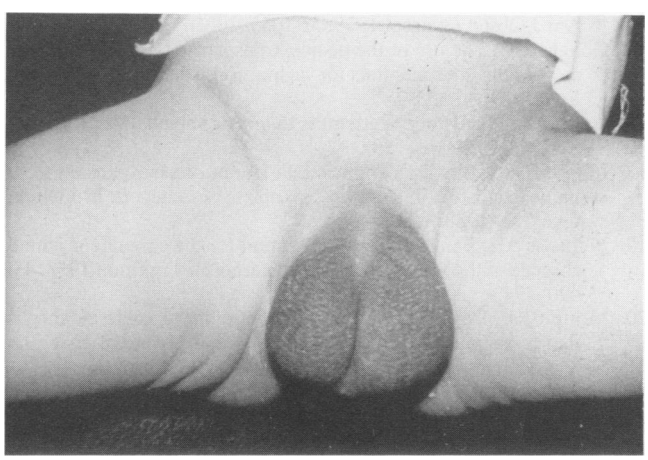


FIG. 11. Congenital absence of the penis in an infant requiring female sex assignment.

Construction of an introitus has brought forth a variety of plastic surgical tissue-moving maneuvers by surgeons working with these patients.[14-24] Some final results are more acceptable than others. In those patients (types A & B) requiring the simpler forms of introitoplasty, excellent feminine appearance and a satisfactory passage for sexual congress can be achieved. The delicate labia minora are difficult to simulate, however, even with the most creative surgical approaches, and the mucus-secreting glands of the normal introitus are congenitally absent in most of these patients.

The pullthrough vaginoplasty to reconstruct patients with distal vaginal atresia and urethrovaginal fistula (Type C) is an elaborate technical exercise involving extensive perineal dissection. The exteriorization of the vagina and, separately, of the urethra is immediately achieved, and the early appearance is satisfactory. Long-term follow-up of this operation, however, has disclosed a number of problems. The first difficulty, found in most of these children, is the unavoidable absence of the mucus apparatus of the normal introitus. The second is the frequent tendency toward stricture formation at the union of the proximal vagina and the external pedicle flaps. The third, and perhaps most objectionable feature is the development of a tangle of hair at the introitus emanating from the inturning flaps of scrotal-like skin. As our results indicate, many of these patients require secondary operations to improve function or appearance.

The complete absence of a vagina necessitates creative surgical reconstruction to simulate a vagina (type D). Although total replacement of the vagina using a sleeve of transposed colon has been used in only two patients in this group, it has proven highly satisfactory in early eval-

uation. The colon is favored over the small bowel because the stretch pain fibers of the smaller diameter small intestine might lead to dyspareunia. The mucus surface of the colon has salutary functional implications for the neovagina. Additionally this operation can be performed early in life; our younger patient was 3 years old at the time of colovaginal replacement.

With respect to evaluating and reporting results of this study, several variables are at play. First is the steady increase in knowledge about the subject matter.[25-27] Understanding of this spectrum of pathology and physiology has been evolving over the four decades since Lawson Wilkins' discovery of the deranged physiology in children born with congenital adrenal hyperplasia.[28] A second obstacle of even greater importance in the reporting of surgical results is the ongoing progress in operative approaches for correcting the deformities that these children present.[29-35] Further, it has become obvious to all workers in this field that sexual function and psychosocial adjustment are the main goals for these patients, rather than simple assessment of their genital appearance.[36-40] It is these subjective factors, so difficult to quantify, that are the ultimate determinants of successful surgical reconstruction.

In this regard, it is difficult to emphasize adequately the compelling need for an ongoing care program encompassing, physical, endocrinologic, and psychologic needs for each patient and each parent caught in the web of the intersex phenomenon. We have learned, in some instances bitterly, the absolute essentiality of a structured, regular, comprehensive program from infancy, through the turbulent adolescent years with its sexual awakenings, and, finally into adult life. This program must have decades-long continuity, and the team must include a pediatrician, an endocrinologist, a psychiatrist or psychologist, and a nurse as well as a surgeon experienced in the vexing surgical problems these fascinating young patients present.

Conclusions

1. Patients with any form of the intersexual malformations benefit from recognition on the first day of life.
2. A comprehensive battery of diagnostic tools is available to pinpoint the pathologic derangement, and to allow accurate sex assignment in a matter of hours, or at the most, days after birth.
3. An effective program of surgical rehabilitation can be undertaken in the early months of life.
4. A program spanning infancy to adulthood must be set up to provide all physical and psychologic needs for the patient and family.
5. Late maladjustment, sexual failure, and psychic distress can largely be avoided today by an informed health care team acting in concert over the patient's lifetime.

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Discussion

DR. BIEMANN OTHERSEN (Charleston, South Carolina): Dr. Ochsner, Dr. Jones, Ladies and Gentlemen, I appreciate the opportunity to discuss this fine presentation and to have reviewed the manuscript. To amass this number of patients with ambiguous genitalia in itself is a real accomplishment. But what comes through in the manuscript is not just meticulous medical and surgical care of these children, but also real compassion and concern for them and their families. This is a stressful situation to every mother and father presented with a child with this problem. I would like to stress the admonition of the authors here about gender assignment and what problems occur when you make the wrong assignment quickly and without a lot of thought and study. As an example, children with exstrophy of the cloaca often have an intussusception of the terminal ileum through the exostrophied bowel, and it looks like a phallus. And you have to address that right away, because if these are boys and you try to raise them as a boy, as they brought out in the paper, it is impossible because of lack of a functional phallus or penis.

I would like to ask a question that may concern a situation that would present itself to any general surgeon who takes care of children with hernia—that is, that is a normal-appearing girl with a hernia and a gonad in the hernia sac. If you open the sac and find the gonad and there is no Fallopian tube, some would advise biopsy of the gonad to make sure this is not testicular feminization or male pseudohermaphroditism. Even though the child looks like a girl, the organ is a testis. If you see a normal Fallopian tube in addition to the gonad, does this mean that the gonad is an ovary or does it mean you should still perform a biopsy on that gonad? Thank you. I enjoyed this presentation very much.

DR. JUDSON RANDOLPH (Closing discussion): Well, Dr. Othersen has raised a good question. If the gonad in a girl's hernia is free of the Fallopian tube, we always biopsy. We take a good, hard look at the others. Sometimes they are odd-looking ovaries, and they might be ovo-testes, or they might even, in the male pseudohermaphrodite, be testes. If there is anything odd, we perform biopsies, but not on every one. Thank you very much for your discussion.